Complete Summary

GUIDELINE TITLE

Task Force on Sudden Cardiac Death of the European Society of Cardiology.

BIBLIOGRAPHIC SOURCE(S)

Priori SG, Aliot E, Blomstrom-Lundqvist C, Bossaert L, Breithardt G, Brugada P, Camm AJ, Cappato R, Cobbe SM, Di Mario C, Maron BJ, McKenna WJ, Pedersen AK, Ravens U, Schwartz PJ, Trusz-Gluza M, Vardas P, Wellens HJ, Zipes DP. Task Force on Sudden Cardiac Death of the European Society of Cardiology. Eur Heart J 2001 Aug;22(16):1374-450. [586 references]

COMPLETE SUMMARY CONTENT

SCOPE

DISEASE/CONDITION(S)

Sudden cardiac death

GUIDELINE CATEGORY

Evaluation
Prevention
Risk Assessment
Treatment

CLINICAL SPECIALTY

Cardiology
Emergency Medicine
Family Practice
Internal Medicine

Nursing
Surgery

INTENDED USERS

Advanced Practice Nurses
Nurses
Physician Assistants
Physicians

GUIDELINE OBJECTIVE(S)

To provide evidence-based recommendations for the prediction and prevention of sudden cardiac death

TARGET POPULATION

Patients at risk for sudden cardiac death

INTERVENTIONS AND PRACTICES CONSIDERED

Risk Stratification for Sudden Cardiac Death Based On:

1. Underlying cardiovascular disease
2. Demographic variables, genetic studies, and personal/family history
3. Clinical, electrocardiographic, echocardiographic markers, including electrophysiologic studies
4. Other risk factors

Primary or Secondary Prophylaxis of Sudden Cardiac Death

1. Angiotensin converting enzyme (ACE) inhibitors
2. Aldosterone receptor blockers
3. Lipid lowering agents
4. Nitrates
5. Magnesium therapy
6. Thrombolytic and antithrombotic therapy, such as aspirin heparin, and oral anticoagulants
7. Revascularization
8. Positive inotropic agents/phosphodiesterase inhibitors
9. Sodium channel blockers, such as lidocaine, encainide, flecainide, and moricizine
10. Other anti-arrhythmic drugs
11. Beta-blockers, such as carvedilol, bisoprolol, and metoprolol
12. Amiodarone
13. Potassium channel blockers, such as d,l sotalol and dofetilide
14. Calcium channel blockers, such as verapamil
15. Implantable cardioverter defibrillators
16. Catheter ablation
17. Surgical ablation
18. Avoidance of exercise and drug triggers

Resuscitation After Cardiac Arrest

1. Basic life support
2. Advanced cardiac life support, including use of automated external defibrillator

MAJOR OUTCOMES CONSIDERED

- Mortality rates (all-cause or total mortality, sudden cardiac death, and arrhythmic death)
- Sensitivity, specificity, and predictive value of risk assessment techniques
- Arrhythmic events

METHODOLOGY

METHODS USED TO COLLECT/SELECT EVIDENCE

Searches of Electronic Databases

DESCRIPTION OF METHODS USED TO COLLECT/SELECT THE EVIDENCE

A literature review was performed through searching databases (Ovid, Medline). A large number of publications in English were reviewed by the committee members during the course of their discussions.

NUMBER OF SOURCE DOCUMENTS

Not stated

METHODS USED TO ASSESS THE QUALITY AND STRENGTH OF THE EVIDENCE

Weighting According to a Rating Scheme (Scheme Given)

RATING SCHEME FOR THE STRENGTH OF THE EVIDENCE

Levels of Evidence:

A. Data derived from multiple randomized clinical trials or meta-analyses
B. Data derived from a single randomized trial or non-randomized studies
C. Consensus opinion of the experts

METHODS USED TO ANALYZE THE EVIDENCE

Review of Published Meta-Analyses
Systematic Review with Evidence Tables

DESCRIPTION OF THE METHODS USED TO ANALYZE THE EVIDENCE

The committee reviewed and ranked the evidence supporting the current recommendations according to companion document to this guideline, the European Society of Cardiology "Recommendations for Task Force Creation and Report Writing" (available from the European Society of Cardiology Web site).

In controversial areas, or on issues without evidence other than usual clinical practice, a consensus was achieved by agreement in the expert panel after thorough deliberations.

METHODS USED TO FORMULATE THE RECOMMENDATIONS

Expert Consensus

DESCRIPTION OF METHODS USED TO FORMULATE THE RECOMMENDATIONS

Not stated

RATING SCHEME FOR THE STRENGTH OF THE RECOMMENDATIONS

Class of Recommendation:

Class I: Condtions for which there is evidence or general agreement that a given procedure or treatment is useful and effective

Class II: Condtions for which there is conflicting evidence or a divergence of opinion about the usefulness/efficacy of a procedure or treatment

Class IIa: Weight of evidence/opinion is in favour of usefulness/efficacy

Class IIb: Usefulness/efficacy is less well established by evidence/opinion

Class III: Conditions for which there is evidence and/or general agreement that the procedure/treatment is not useful/effective and in some cases may be harmful

COST ANALYSIS

A formal cost analysis was not performed and published cost analyses were not reviewed.

METHOD OF GUIDELINE VALIDATION

External Peer Review
Internal Peer Review

DESCRIPTION OF METHOD OF GUIDELINE VALIDATION

This document was reviewed by the Committee for Practice Guidelines and Policy Conferences and by external reviewers.

RECOMMENDATIONS

MAJOR RECOMMENDATIONS

Note from the National Guideline Clearinghouse (NGC): The European Society of Cardiology (ESC) has updated two sections of this guideline. The updated recommendations are available at the ESC Web site. NGC will update this summary when the updated version of the full text document is published.

The class of recommendations (I-III) and level of evidence (A-C) are defined at the end of the "Major Recommendations" field.

Recommendations for Risk Stratification for Sudden Cardiac Death: Myocardial Infarction and Heart Failure

Class I:

- Demographic variables (Level of Evidence A; Yap et al., 2000)
- Left ventricular ejection fraction (Level of Evidence A; Yap et al., 2000; Stevenson & Ridker 1996; Copie et al., 1996)
- Heart rate variability or baroreflex sensitivity (Level of Evidence A; La Rovere et al., 1998; Zuanetti et al., 1996; Malik et al., 2000)
- Left ventricular volume (Level of Evidence A; Nicolosi et al., 1996)

Class IIa:

- Ventricular premature beats (Level of Evidence A; Statters et al., 1996; Maggioni et al., 1993)
- Non-sustained ventricular tachycardia (Level of Evidence A; Hohnloser et al., 1999; Mushlin et al., 1998; Buxton et al., 1999)
- Resting heart rate (Level of Evidence A; Maggioni et al., 1993)

Class IIb:

- Late potential (Level of Evidence A; Farrell et al., 1991; McClements & Adgey, 1993; Steinberg et al., 1992; Ikeda et al., 2000)
- QT interval (Level of Evidence B; Schwartz & Wolf, 1978)
- Electrophysiological study (Level of Evidence A; Peterson, Shaw, & Califf, 1997; Wellens et al., 1997; Caruso et al., 1997; Andresen et al., 1999)
- T wave alternans (Level of Evidence B; Hohnloser et al., 1999; Ikeda et al., 2000)
- Heart rate turbulence (Level of Evidence B; Schmidt et al., 1999)
- Patency of infarct-related artery (Level of Evidence B; Hohnloser et al., 1994; de Chillou et al., 1997)

Class III:

- QT dispersion (Level of Evidence B; Zabel et al., 1998)

Recommendations for Primary Prevention* of Sudden Cardiac Death: Drugs Without Electrophysiological Properties in Patients With Heart Failure

Class I:

- Beta-blockers (Level of Evidence A; "Effect of metoprolol CR/XL", 1999; "The Cardiac Insuffciency Bisoprolol Study II (CIBIS-II)", 1999; Furberg, Hawkins, & Lichstein, 1984; Packer et al., 1996; "The Beta-Blocker Pooling Project (BBPP)", 1988; Kendall et al., 1995; Gottlieb, McCarter, & Vogel, 1998)
- Angiotensin converting enzyme inhibitors (Level of Evidence A; "Effect of enalapril on survival in patients with reduced left ventricular ejection fractions and congestive heart failure.", 1991; Hennekens et al., 1996; Ambrosioni, Borghi, & Magnani, 1995; "ISIS-4: a randomised factorial trial", 1995; "GISSI-3: effects of lisinopril and transdermal glyceryl trinitrate", 1994; "Effect of enalapril on mortality and the development of heart failure", 1992; "Effects of enalapril on mortality in severe congestive heart failure", 1987).
- Aldosterone receptor blockers (Level of Evidence B; Pitt, Zannad, & Remme, 1999)

Class III:

- Positive inotropic agents phosphodiesterase inhibitors (Level of Evidence B; Packer et al., 1991)
- Digoxin (Level of Evidence B; "The effect of digoxin on mortality and morbidity in patients with heart failure.", 1997)

*Primary prevention refers to prevention of sudden cardiac death in patients who had not experienced sustained ventricular tachyarrhythmias prior to therapy.

Recommendations for Primary Prevention of Sudden Cardiac Death: Drugs Without Electrophysiological Properties During and After Myocardial Infarction*

Class I:

- Beta-blockers (Level of Evidence A; "Effect of metoprolol CR/XL", 1999; "The Cardiac Insuffciency Bisoprolol Study II (CIBIS-II)", 1999; Furberg, Hawkins, & Lichstein, 1984; "The Beta-Blocker Pooling Project (BBPP)", 1988; Kendall et al., 1995; Gottlieb, McCarter, & Vogel, 1998)
- Angiotensin converting enzyme inhibitors (Level of Evidence B; "Effect of enalapril on survival in patients with reduced left ventricular ejection fractions and congestive heart failure.", 1991; Ambrosioni, Borghi, & Magnani, 1995; "ISIS-4: a randomised factorial trial", 1995; "GISSI-3: effects of lisinopril and transdermal glyceryl trinitrate", 1994; "Effect of enalapril on mortality and the development of heart failure", 1992; "Effects of enalapril on mortality in severe congestive heart failure.", 1987)
- Lipid lowering agents (Level of Evidence A; Ambrosioni, Borghi, & Magnani, 1995; "Randomised trial of cholesterol lowering in 4444 patients with coronary heart disease", 1994; Sacks et al., 1996; "Prevention of cardiovascular events and death", 1998)

Class IIa:

- Poly-unsaturated fatty acids (eicosapentaenoic acid+docosahexaenoic acid) (Level of Evidence B; "Dietary supplementation with n-3 polyunsaturated fatty acids", 1999; Marchioli & Valagussa, 2000)

Class III:

- Nitrates (Level of Evidence A; Ambrosioni, Borghi, & Magnani, 1995; "ISIS-4: a randomised factorial trial", 1995; "GISSI-3: effects of lisinopril and transdermal glyceryl trinitrate", 1994; Yusuf et al., 1988)
- Magnesium (Level of Evidence A; Teo & Yusuf, 1993; Woods & Fletcher, 1994; Antman, 1995)

*Only drugs evaluated in studies with sudden cardiac death or arrhythmic death as primary or secondary end-points are considered above, together with interventions that failed to demonstrate survival benefit or that worsened survival. As discussed in the original guideline, revascularization, antithrombotic, anticoagulants reduce total mortality and are therefore pivotal treatments.

Recommendations for Primary Prevention of Sudden Cardiac Death: Drugs With Electrophysiological Properties in Post-Myocardial Infarction Patients*

Class IIa:

- Amiodarone** (Level of Evidence A***; "Effect of prophylactic amiodarone on mortality", 1997; Julian et al., 1997; Cairns et al., 1997; Elizari et al., 2000; Ceremuzynski et al., 1992; Burkart et al., 1990; Singh et al., 1995)

Class III:

- Potassium channel blockers:
    - d-sotalol (Level of Evidence B****; Waldo et al., 1996)
    - dofetilide (Level of Evidence A; Torp-Pedersen et al., 1999; Kober et al., 2000)
- Calcium channel blockers (Level of Evidence B; Teo, Yusuf, & Furberg, 1993; Held, Yusuf, & Furberg, 1989; "Effect of verapamil on mortality and major events", 1990; Waters et al., 1990; Yusuf, Held, & Furberg, 1991)
- Sodium channel blockers (Level of Evidence B; "Preliminary report: effect of encainide and flecainide", 1989; MacMahon et al., 1988; Teo, Yusuf, & Furberg, 1993; "Effect of the antiarrhythmic agent moricizine", 1992; Echt et al., 1991)

*Beta-blockers have been included previously (drugs without electrophysiological properties.

**Data refer also to patients with congestive heart failure.

***Reduces sudden cardiac death, reduction of total mortality showed borderline significant reduction.

****Worsens prognosis.

Recommendations for the Use of an Implantable Defibrillator for Primary Prevention in Patients at Risk of Sudden Cardiac Death According to Specific Underlying Clinical Conditions

Class I:

- Post-myocardial infarction, ejection fraction <40%, clinical non-sustained ventricular tachycardia, sustained ventricular tachycardia at programmed electrical stimulation (Level of Evidence B*; Buxton et al., 1999; Moss et al., 1996)

Class III:

- Post- myocardial infarction, ejection fraction <36%, late potentials, indication for coronary artery bypass graft (Level of Evidence B; Bigger, 1997)

*Data collected in post myocardial infarction patients with stables ischaemic heart disease, and might not apply to patients with recent myocardial infarction (<3 weeks).

Recommendations for the Use of Drugs With Electrophysiological Properties, Implantable Defibrillators, Catheter Ablation and Surgery for Secondary Prevention in Post Myocardial Infarction Patients With Documented Sustained Ventricular Arrhythmias According to Their Clinical Presentation

Note: Secondary prevention refers to prevention of sudden cardiac death in patients who had already experienced a sustained ventricular tachyarrhythmia prior to the onset of therapy.

Resuscitated Ventricular Tachycardia/Ventricular Fibrillation, Spontaneous Sustained Haemodynamically Non-tolerated Ventricular Tachycardia

Class I:

- Implantable defibrillator (Level of Evidence A; "A comparison of antiarrhythmic-drug therapy", 1997; Kuck et al., 2000)

Class IIa:

- Beta-blockers (Level of Evidence C*; Kuck et al., 2000)
- Amiodarone (Level of Evidence C*; "Randomized antiarrhythmic drug therapy", 1993; "A comparison of antiarrhythmic-drug therapy", 1997; Connolly et al., 2000; Kuck et al., 2000)

Class III:

- Potassium channel blockers (Level of Evidence C; Haverkamp et al., 1997; Bocker et al., 1996)
- Calcium channel blockers (Opinion of the Task Force Panel)
- Sodium channel blockers (Level of Evidence C; Mason, 1993; Kuck et al., 2000)

Spontaneous, Sustained, Well Tolerated, Monomorphic Ventricular Tachycardia

Class IIa:

- Beta-blockers (Opinion of the Task Force Panel**)
- Amiodarone (Opinion of the Task Force Panel**)

Class IIb:

- Implantable defibrillator (Level of Evidence B; Raitt et al., 2001)
- Ablation (Level of Evidence C**; Morady et al., 1993; Stevenson et al., 1993)
- Surgery (Level of Evidence C; van Hemel et al., 1989; van Hemel et al., 1996)

Class III:

- Potassium channel blockers (Opinion of the Task Force Panel)
- Calcium channel blockers (Opinion of the Task Force Panel)
- Sodium channel blockers (Opinion of the Task Force Panel)

*As an alternative to implantable defibrillators when the device in not implanted.

**Possibly reduces recurrence, unlikely to reduce sudden cardiac death.

Recommendations for Risk Stratification for Sudden Cardiac Death: Hypertrophic Cardiomyopathy

Class I:

- Cardiac arrest (or sustained ventricular tachycardia) (Level of Evidence B; Cecchi, Maron, & Epstein, 1989; Elliott et al., 1999; Maron et al., 2000)

Class IIa:

- Family history of sudden death (Level of Evidence B; Spirito et al., 1997; Maron, 1997; Maron et al., 1999; Redwood, Moolman-Smook, & Watkins, 1999; Elliott et al., 2000)
- Syncope* (Level of Evidence B; McKenna et al., 1981; McKenna et al., 1988; McKenna & Deanfield, 1984; Elliott et al., 2001; Elliott et al., 2000; Fananapazir et al., 1992)
- Extreme left ventricular hypertrophy (>3 cm maximum wall thickness)** (Level of Evidence B; Maron, Roberts, & Epstein, 1982; Spirito et al., 2000; Elliott et al., 2001; Elliott et al., 2000)

- Hypotensive blood pressure response to exercise (Level of Evidence B; Elliott et al., 2001; Elliott et al., 2000; Varnava et al., 1999; Frenneaux et al., 1990; Sadoul et al., 1997; Counihan et al., 1991; Olivotto et al., 1999)
- Non-sustained ventricular tachycardia (Holter) (Level of Evidence B; McKenna et al., 1988; Elliott et al., 2001; Elliott et al., 2000; McKenna et al., 1981; Maron et al., 1981; Spirito et al., 1994; McKenna et al., 1994; Fananapazir et al., 1992; Maron et al., 2000)

Class IIb:

- High-risk mutations (Level of Evidence B; Spirito et al., 1997; Maron, 1997; Redwood, Moolman-Smook, & Watkins, 1999; Watkins et al., 1995; Varnava et al., 1999)

Class III:

- Inducible ventricular arrhythmias at programmed electrical stimulation (Level of Evidence C; Fananapazir et al., 1992; Cecchi, Maron, & Epstein, 1989; Elliott et al., 1999; Maron et al., 2000; Kuck et al., 1988; Saumarez et al., 1995)
- Left ventricular outflow gradient (Level of Evidence B; McKenna et al., 1981)
- Mitral regurgitation (moderate-severe) (Level of Evidence C; McKenna et al., 1981)
- Chest pain/dyspnoea (Level of Evidence C; McKenna et al., 1981)
- Paroxysmal atrial fibrillation (Level of Evidence B; Stafford et al., 1986)

*Risk appears to be greatest when repetitive, or associated with exertion, or in children.

**Usually ventricular septum.

Recommendations for Prevention of Sudden Cardiac Death: Hypertrophic Cardiomyopathy

Primary Prevention

Class IIa:

- Implantable cardioverter defibrillator (Level of Evidence B; Cecchi, Maron, & Epstein, 1989; Maron et al., 2000)

Class IIb:

- Amiodarone (Level of Evidence B; McKenna et al., 1985; Cecchi et al., 1998)

Class III:

- Beta-blockers (Level of Evidence C; Spirito et al., 1997; Maron, 1997; Wigle et al., 1985; McKenna et al., 1981)
- Verapamil (Level of Evidence C; Spirito et al., 1997; Maron, 1997; Wigle et al., 1985)

Secondary Prevention

Class I:

- Implantable cardioverter defibrillator (Level of Evidence B; Cecchi, Maron, & Epstein, 1989; Maron et al., 2000)

Recommendations for Risk Stratification for Sudden Cardiac Death: Right Ventricular Cardiomyopathy

Class IIa

- Diffuse right ventricular dilatation (Level of Evidence C; Peters, 1997; Corrado et al., 1997; Peters, 1995)
- Left ventricular involvement (Level of Evidence C; Peters, 1997; Corrado et al., 1997; Peters, 1995)
- Right ventricular dysfunction/dilatation + inducible sustained ventricular tachycardia (Level of Evidence C; Peters, 1997; Corrado et al., 1997; Peters, 1995)
- Previous cardiac arrest/ventricular fibrillation (Level of Evidence C; Peters, 1997; Corrado et al., 1997; Peters, 1995)

Class IIb:

- Family history of arrhythmogenic right ventricular cardiomyopathy and sudden cardiac death (Level of Evidence C; Daliento et al., 1995; Blomstrom-Lundqvist, Sabel, & Olsson, 1987; Marcus et al., 1989; Corrado et al., 1997; Turrini et al., 1999)
- Syncope (Level of Evidence C; Daliento et al., 1995; Blomstrom-Lundqvist, Sabel, & Olsson, 1987; Marcus et al., 1989; Corrado et al., 1997; Turrini et al., 1999)
- Late potentials + right ventricular dysfunction (Level of Evidence C; Daliento et al., 1995; Blomstrom-Lundqvist, Sabel, & Olsson, 1987; Marcus et al., 1989; Corrado et al., 1997; Turrini et al., 1999; Leclercq & Coumel, 1993; Mehta et al., 1996; Hermida et al., 1997)
- Ventricular tachycardia (Level of Evidence C; Daliento et al., 1995; Blomstrom-Lundqvist, Sabel, & Olsson, 1987; Marcus et al., 1989; Corrado et al., 1997; Turrini et al., 1999; Lemery et al., 1989; Leclercq & Coumel, 1989; Leclercq et al., 1996)
- Programmed electrical stimulation* (Opinion of the Task Force Panel)

Class III:

- QT dispersion and T wave complexity (Level of Evidence C; Benn, Hansen, & Pedersen, 1999; De Ambroggi et al., 1997)
- Premature ventricular contractions (Level of Evidence C; Benn, Hansen, & Pedersen, 1999; De Ambroggi et al., 1997; Leclercq & Coumel, 1989)

*Programmed electrical stimulation inducibility identifies patients at high risk of sudden cardiac death when associated with enlargement and right ventricular dysfunction.

Recommendations for Prevention of Sudden Cardiac Death: Right Ventricular Cardiomyopathy--Ventricular Tachycardia

Primary Prevention

Class IIa:

- Implantable cardioverter defibrillator (Opinion of the Task Force Panel)

Class IIb

- Antiarrhythmic agent drugs* (Level of Evidence C; Berder et al., 1995; Wichter et al., 1992; Lemery et al., 1989; Leclercq & Coumel, 1989)

Class III:

- Right frequency ablation (Level of Evidence C; Harada et al., 1998; Ellison et al., 1998)

Class III:

- Asymptomatic-high risk
- Antiarrhythmic agent drugs* (Level of Evidence C; Berder et al., 1995; Lemery et al., 1989; Leclercq & Coumel, 1989)
- Implantable cardioverter defibrillator (Level of Evidence C; Link et al., 1997; Breithardt et al., 1994)

Secondary Prevention

Class I:

- Implantable cardioverter defibrillator (Level of Evidence C; Link et al., 1997; Breithardt et al., 1994)

Class III:

- Antiarrhythmic agent drugs (Level of Evidence C; Berder et al., 1995; Wichter et al., 1992; Lemery et al., 1989; Leclercq & Coumel, 1989)
- Right frequency ablation (Level of Evidence C; Harada et al., 1998; Ellison et al., 1998)

*Data on antiarrhythmic agent drugs are mainly retrospective analysis. In one prospective study sotalol treatment guided by acute oral drug testing during electrophysiological study demonstrated higher efficacy than other agents.

Recommendations for Risk Stratification for Sudden Cardiac Death: Dilated Cardiomyopathy

Class I:

- Previous cardiac arrest/ventricular fibrillation (Level of Evidence B; Luu et al., 1989; Kelly & Coats, 1997; Tamburro & Wilber, 1992)
- Sustained ventricular tachycardia (Level of Evidence B; Brembilla-Perrot et al., 1991; Middlekauff et al., 1993; Knight et al., 1999; Meinertz et al., 1984)

Class IIa:

- Syncope (Level of Evidence B; "Effect of metoprolol CR/XL", 1999; Fuster et al., 1981; Di Lenarda et al., 1994; Dec & Fuster, 1994; Stewart, McKenna, & Oakley, 1990; Hofmann et al., 1988; Romeo et al., 1989; Gradman et al., 1989; Middlekauff et al., 1993; Knight et al., 1999)

Class IIb:

- Ejection fraction (Level of Evidence B; "Effect of metoprolol CR/XL", 1999; Fuster et al., 1981; Di Lenarda et al., 1994; Dec & Fuster, 1994; Stewart, McKenna, & Oakley, 1990; Hofmann et al., 1988; Romeo et al., 1989; Gradman et al., 1989; Keogh, Baron, & Hickie, 1990)
- Non sustained ventricular tachycardia (Level of Evidence B; Stewart, McKenna, & Oakley, 1990; Hofmann et al., 1988; Kron et al., 1988; Meinertz et al., 1984; Wilber, 1996)

Class III:

- Inducibility at programmed electrical stimulation (Level of Evidence B; Middlekauff et al., 1993; Knight et al., 1999; Kron et al., 1988; Meinertz et al., 1984)

Recommendations for Prevention of Sudden Cardiac Death: Dilated Cardiomyopathy

Class I

- Angiotensin converting inhibitors (Level of Evidence B; Hennekens et al., 1996)
- Beta-blocker (Level of Evidence B; "The Cardiac Insuffciency Bisoprolol Study II (CIBIS-II): a randomised trial.", 1999)
- Implantable cardioverter defibrillator (secondary prevention) (Opinion of Task Force Panel)

Class IIa

- Implantable cardioverter defibrillator (primary prevention) (Level of Evidence B; Connolly et al., 2000; Steinberg, Ehlert, & Cannon, 1997)
- Aldosterone receptor blockers (Level of Evidence B; Pitt, Zannad, & Remme, 1999)

Class IIb:

- Amiodarone (Level of Evidence B; Singh et al., 1995; Doval et al., 1994; Connolly et al., 2000)

Recommendations for Risk Stratification Sudden Cardiac Death: Long QT Syndrome

Class I:

- Syncope (Level of Evidence B; Schwartz, Priori, & Napolitano, 2000; Moss et al., 1991)
- Torsades de Pointes/Ventricular fibrillation/Cardiac arrest (Level of Evidence B; Schwartz, Priori, & Napolitano, 2000; Moss et al., 1991)
- JLN recessive variant (Level of Evidence B; Schwartz, Priori, & Napolitano, 2000; Moss et al., 1991; Schwartz et al., 2001)
- LQT3 genetic variant (Level of Evidence C; Schwartz, Priori, & Napolitano, 2000; Zareba et al., 1998; Schwartz et al., 2001)

Class IIa

- Rate corrected QT interval >600 msec (Level of Evidence C; Moss et al., 1991)
- Cardiac events in infants (Opinion of the Task Force Panel)
- Post partum period (Level of Evidence C; Rashba et al., 1998)
- Female gender (Level of Evidence C; Locati et al., 1998)
- Syndactyly and atrioventricular block (Level of Evidence C; Marks, Trippel, & Keating, 1995)
- T wave alternans (macroscopic) (Level of Evidence C; Zareba et al., 1994)

Class IIb:

- Family history (Opinion of the Task Force Panel)
- QT dispersion (Level of Evidence C; Priori et al., 1994)

Class III:

- Programmed electrical stimulation (Level of Evidence C; Bhandari et al., 1985)

Recommendations for Prevention of Sudden Cardiac Death: Long QT Syndrome

Primary Prevention

<u>Avoidance of QT Prolonging Agents/K+ Lowering Agents</u>

Class IIa:

- Symptomatic (Level of Evidence C; Schwartz, Priori, & Napolitano, 2000)
- Silent gene carriers (Level of Evidence C; Schwartz, Priori, & Napolitano, 2000; Napolitano et al., 2000)
- Asymptomatic (Level of Evidence C; Schwartz, Priori, & Napolitano, 2000; Napolitano et al., 2000)

<u>Avoidance of Competitive Sports/Strenuous Activity</u>

Class I:

- Symptomatic (Level of Evidence C; Schwartz, Priori, & Napolitano, 2000)

Class IIa:

- Silent gene carriers (Level of Evidence C; Schwartz, Priori, & Napolitano, 2000)
- Asymptomatic (Level of Evidence C; Schwartz, Priori, & Napolitano, 2000)

Beta-blockers

Class I:

- Symptomatic (Level of Evidence B; Moss et al., 2000)

Class IIa

- Asymptomatic (Level of Evidence C; Moss et al., 2000)

Left Cardiac Sympathetic Denervation+Beta-Blockers

Class IIb:

- Symptomatic with recurrences on beta-blockers (Level of Evidence B; Schwartz et al., 1991)

Pacemaker (Plus Beta-Blockers)

Class IIb:

- Symptomatic with pause- or bradycardia-dependent arrhythmias (Level of Evidence C; Moss et al., 1991; Dorostkar et al., 1999)

Implantable Cardioverter Defibrillator+Beta-Blockers

Class IIa:

- Symptomatic with recurrences on beta-blockers (Level of Evidence C; Schwartz, Priori, & Napolitano, 2000; Zareba et al., 1998)

Secondary Prevention

Class I:

- Implantable cardioverter defibrillator +beta-blockers (Level of Evidence C; Schwartz, Priori, & Napolitano, 2000; Moss et al., 2000)
- Avoidance of competitive sport/strenuous activity* (Level of Evidence C; Schwartz, Priori, & Napolitano, 2000)

- Avoidance of QT prolonging agents* (Level of Evidence C; Schwartz, Priori, & Napolitano, 2000)

  *Life-style measures to be adopted in conjunction with implantable cardioverter defibrillator implant in cardiac arrest survivors.

Recommendations for Risk Stratification for Sudden Cardiac Death: Brugada Syndrome

Class IIa:

- Family history for sudden cardiac death (Level of Evidence C; Brugada, Brugada, & Brugada, 1998)
- Syncope (Level of Evidence C; Brugada, Brugada, & Brugada, 1998; Priori et al., 2000)

Class IIb:

- Ventricular tachycardia/ventricular fibrillation inducibility (Level of Evidence C; Brugada, Brugada, & Brugada, 1998; Priori et al., 2000)

Recommendations for Prevention of Sudden Cardiac Death: Brugada Syndrome

Primary Prevention

Class I:

- Implantable cardioverter defibrillator
- Symptomatic for syncope/ventricular tachycardia (Level of Evidence B; Brugada, Brugada, & Brugada, 1998; Priori et al., 2000)

Class IIb:

- Asymptomatic with inducible ventricular tachycardia /ventricular fibrillation (Level of Evidence C; Brugada, Brugada, & Brugada, 1998; Priori et al., 2000)

Class III:

- Asymptomatic with non-inducible ventricular tachycardia/ventricular fibrillation (Level of Evidence C; Brugada, Brugada, & Brugada, 1998; Priori et al., 2000)
- Antiarrhythmic agent drugs (Level of Evidence C; Brugada, Brugada, & Brugada, 1998; Priori et al., 2000)

Secondary Prevention

Class I:

- Implantable cardioverter defibrillator (Level of Evidence B; Brugada, Brugada, & Brugada, 1998; Priori et al., 2000)

Class III:

- Antiarrhythmic agent drugs (Level of Evidence C; Brugada, Brugada, & Brugada, 1998; Priori et al., 2000)

Recommendations for Risk Stratification for Sudden Cardiac Death: Catecholaminergic Polymorphic Ventricular Tachycardia

Class I

- Documented ventricular fibrillation (Level of Evidence C; Leenhardt et al., 1995)

Class IIa

- Family history of sudden cardiac death (Level of Evidence C; Leenhardt et al., 1995)
- Early onset of symptoms (paediatric age) (Level of Evidence C; Leenhardt et al., 1995)

Class IIb:

- Syncope (Level of Evidence C; Leenhardt et al., 1995)

Recommendations for Prevention of Sudden Cardiac Death: Catecholaminergic Polymorphic Ventricular Tachycardia

Primary Prevention

Class IIa:

- Beta-blockers (Level of Evidence C; Leenhardt et al., 1995)

Class IIb:

- Implantable cardioverter defibrillator (Opinion of the Task Force Panel)

Secondary Prevention

Class I:

- Implantable cardioverter defibrillator (plus beta-blockers) (Opinion of the Task Force Panel)

Class IIa:

- Beta-blockers (Level of Evidence C; Leenhardt et al., 1995)

Recommendations for Risk Stratification for Sudden Cardiac Death: Aortic Stenosis

Before Aortic Valve Replacement

Class I:

- Symptomatic patients (Level of Evidence C; Ross & Braunwald, 1968; Otto et al., 1997; Horstkotte & Loogen, 1988; Pellikka et al., 1990; Lund et al., 1996)
- Asymptomatic patients

Class IIa:

- Significant ventricular arrhythmia (patients presenting with ventricular tachycardias inducible at programmed electrical stimulation) (Level of Evidence C; Martinez-Rubio et al., 1997)
- Left ventricular dysfunction (Level of Evidence C; Braunwald, 1993)
- Abnormal response to exercise (Level of Evidence C; Braunwald, 1993)

Class IIb:

- Degree of stenosis (Level of Evidence C; Otto et al., 1997)

After Aortic Valve Replacement

Class IIa:

- Significant ventricular arrhythmia (Level of Evidence C; Martinez-Rubio et al., 1997)

Recommendations for Prevention of Sudden Cardiac Death in Aortic Stenosis

Class I:

- Implantable cardioverter defibrillator (secondary prevention) (Level of Evidence C; Martinez-Rubio et al., 1997)
- Aortic valve replacement (Level of Evidence B; Pellikka et al., 1990; Czer et al., 1988)

Class IIa:

- Amiodarone (Opinion of the Task Force Panel)

Recommendations for Risk Stratification for Sudden Cardiac Death: Mitral Valve Prolapse

Class I:

- Cardiac arrest or ventricular tachycardias (Level of Evidence C; Vohra et al., 1993; Boudoulas et al., 1990)

Class IIa:

- Leaflet redundancy/mixomatous valve (Level of Evidence C; Nishimura et al., 1985)
- Family history of sudden death (Level of Evidence C; Vohra et al., 1993; Boudoulas et al., 1990)

Class IIb:

- QT interval and QT dispersion (Level of Evidence C; Tieleman et al., 1995; Kulan et al., 1996)
- Frequent and complex ventricular arrhythmia (Level of Evidence C; Savage et al., 1983)
- Mitral valve regurgitation (Level of Evidence C; Zuppiroli et al., 1995; Nishimura et al., 1985; Boudoulas et al., 1990)
- Programmed electrical stimulation inducibility (Level of Evidence C; Babuty et al., 1994)
- Signal Averaged electrocardiogram (Level of Evidence C; Babuty et al., 1994)

Recommendations for Prevention of Sudden Cardiac Death: Mitral Valve Prolapse

Class I:

- Implantable cardioverter defibrillator (secondary prevention) (Opinion of the Task Force Panel)

Class III:

- Beta-blockers* (Opinion of the Task Force Panel)

*Beta-blockers may be useful in reducing premature ventricular contractions but their value in preventing sudden cardiac death has not been demonstrated.

Recommendations for Risk Stratification for Sudden Cardiac Death: Anomalous Origin of Coronary Arteries

Class I:

- Cardiac arrest in history (Level of Evidence C; Kimbiris et al., 1978; Liberthson, Dinsmore, & Fallon, 1979; Cheitlin, De Castro, & McAllister, 1974; Viskin et al., 1992)

Class IIa:

- Young patients, especially males with exertional chest pain or syncope and electrocardiogram changes (Level of Evidence C; Kimbiris et al., 1978;

Liberthson, Dinsmore, & Fallon, 1979; Cheitlin, De Castro, & McAllister, 1974; Viskin et al., 1992)

Recommendations for Prevention of Sudden Cardiac Death: Anomalous Origin of Coronary Arteries

Class I:

- Surgery (Level of Evidence C; Kimbiris et al., 1978; Liberthson, Dinsmore, & Fallon, 1979; Cheitlin, De Castro, & McAllister, 1974)

Recommendations for Risk Stratification for Sudden Cardiac Death in Myocardial Bridging

Class I:

- Cardiac arrest or symptomatic ventricular tachycardia in history (Level of Evidence C; Desseigne, Tabib, & Loire, 1991; Cutler & Wallace, 1997)

Class IIa

- Proven myocardial ischaemia (Level of Evidence C; Tio et al., 1997)

Recommendations for Prevention of Sudden Cardiac Death: Myocardial Bridging

Class I:

- Surgery in ischaemic patients (Level of Evidence C; Stables et al., 1995)

Class IIa:

- Beta-blockers (Level of Evidence C; Schwarz et al., 1996)

Class III

- Nitrates (Level of Evidence C; Ge et al., 1999)

Recommendations for Risk Stratification for Sudden Cardiac Death: Wolff-Parkinson-White Syndrome

Class IIa

- Short (<250 ms) RR interval during atrial fibrillation (Level of Evidence B; Klein et al., 1979; Attoyan et al., 1994; Montoya et al., 1991; Auricchio et al., 1991)
- Short (<270 ms) anterograde refractory period of the accessory pathway (Level of Evidence B; Timmermans et al., 1995; Munger et al., 1993; Montoya et al., 1991)

- Multiple accessory pathways (Level of Evidence C; Timmermans et al., 1995; Munger et al., 1993; Attoyan et al., 1994; Montoya et al., 1991)

Class IIb:

- Loss of pre-excitation ajmaline or procainamide test (lower risk) (Level of Evidence C; Wellens et al., 1980)

Class III:

- Syncope (Level of Evidence C; Auricchio et al., 1991)

Recommendation for Prevention of Sudden Cardiac Death: Wolff-Parkinson-White Syndrome

Class I:

- Catheter ablation-secondary prevention (Level of Evidence C; Zipes et al., 1995)
- Catheter ablation: symptomatic patients with atrial fibrillation and rapid response via the accessory pathway (Level of Evidence C; Zipes et al., 1995)

Class IIa:

- Catheter ablation: asymptomatic patients with family history of sudden cardiac death, high risk professions, athletes (Level of Evidence C; Zipes et al., 1995)

Class IIb:

- Amiodarone, class Ia, Ic antiarrhythmic drugs* (Opinion of the Task Force Panel)

*Alternative to ablation in asymptomatic patients.

Recommendations for Risk Stratification for Sudden Cardiac Death: Conduction System Abnormalities

Acquired Atrioventricular Block in Adults

Class IIa:

- III degree atrioventricular block (Level of Evidence C; Edhag & Swahn, 1976; Hindman et al., 1978)
- II degree atrioventricular block type II (Level of Evidence C; Shaw et al., 1985)
- Syncope (Level of Evidence B; McAnulty et al., 1982; Scheinman et al., 1982)
- Coexistent heart disease or heart failure (Level of Evidence C; Hindman et al., 1978)

Congenital III Degree Atrioventricular Block

Class I:

- Syncope (Level of Evidence B; Michaelsson, Jonzon, & Riesenfeld, 1995; Michaelsson & Engle, 1972)
- Prolonged rate corrected QT interval (Level of Evidence B; Solti et al., 1992; Silka et al., 1990)
- Congenital heart disease (Level of Evidence C; Solti et al., 1992; Silka et al., 1990)

Chronic Bifascicular and Trifascicular Block

Class I:

- Coexistent heart disease or heart failure (Level of Evidence B; Dhingra et al., 1978)

Class IIa:

- Syncope (Level of Evidence B; Morady et al., 1984; Dhingra et al., 1979; Click et al., 1987; Scheinman et al., 1982)
- His-Ventricle (HV) $\geq$100 ms or pacing induced infra-His block (Level of Evidence C; Scheinman et al., 1974; Tonkin, Heddle, & Tornos, 1978)
- Programmed electrical stimulation inducibility (Level of Evidence B; Ezri et al., 1983; Morady et al., 1984)

Recommendations for Prevention of Sudden Cardiac Death: Conduction System Abnormalities

Class I:

- Pacemaker in higher risk subgroups (Level of Evidence C; Shaw et al., 1985; Johansson, 1966; Strasberg et al., 1981; Michaelsson, Jonzon, & Riesenfeld, 1995; Michaelsson & Engle, 1972)

Recommendations for Risk Stratification for Sudden Cardiac Death in Young Athletes*

Class I:

- 12-lead electrocardiogram (Level of Evidence A; Pelliccia & Maron, 1995)

Class IIa:

- Physical examination (Level of Evidence B; Corrado et al., 1998; Pelliccia & Maron, 1995)
- Personal history (Level of Evidence B; Maron et al., 1996; Glover & Maron, 1998; Pfister, Puffer, & Maron, 2000)

Class IIb:

- Family history (Level of Evidence B; Maron et al., 1996; Glover & Maron, 1998; Pfister, Puffer, & Maron, 2000)

*These recommendations are offered solely in a scientific context, without particular consideration to other important issues concerning cardiovascular screening, such as implementation and cost efficacy. Indeed, the guideline developers understand that some European countries may not wish to formulate screening in the design offered here and that the inclusion of a 12-lead electrocardiogram to the protocol may not be regarded as feasible.

Guidelines for the Use of Autonomic External Defibrillators

- In cases of sudden cardiac arrest, early defibrillation by the first responding professional rescuer is now well accepted as the standard of care. In the international 2000 guidelines for the use of autonomic external defibrillators by emergency medical service providers and first responders it is recommended that:
    - every ambulance, which might respond to a cardiac arrest, must carry a defibrillator with personnel trained permitted to use it
    - defibrillation should be one of the core competencies of doctors, nurses and other health care professionals
    - defibrillators should be widely placed on general hospital wards
    - to investigate the feasibility and efficacy of allowing all those assigned to the management of cardiac arrest in the community to be trained and permitted to defibrillate. Refresher training should be carried out at least every 6 months. Specifically certified instructors working within a medically controlled system should give the training.
- The medical profession is urged to increase awareness of the public, of those responsible for emergency medical services and of those with regulatory powers, to permit changes in practice and legislation where necessary.
- It is essential to integrate the concept of early defibrillation into an effective emergency cardiac care system, which includes early access to the emergency medical services system, early cardiopulmonary resuscitation by the first witness, early defibrillation when indicated, and early advanced care.
- All emergency personnel, should be trained and permitted to operate a defibrillator if their professional activities require that they respond to persons with cardiac arrest. This includes all first responding emergency personnel working in an organized emergency medical service system, both in and outside the hospital.
- All emergency ambulances that respond to or transport cardiac patients should be equipped with a defibrillator.
- Defibrillation should be a core competence of all health care professionals including nurses, and defibrillators should be widely available on general hospital wards.
- All defibrillator programs must operate within medical control by qualified and experienced physicians. They should ensure that every link of the chain of survival is in place and should have access to all information required to permit system audit.
- To monitor the program, there must be appropriate registration of the interventions according to the Utstein style.

Recommendations for the Use of Autonomic External Defibrillators in the Prevention of Sudden Cardiac Death

Class I:

- Use by emergency medical service personnel (Level of Evidence B; Herlitz et al., 1999)
- Use by police (Level of Evidence C; White et al., 1996; Mosesso et al., 1998)
- Use in commercial aircraft (Level of Evidence B; O´Rourke, Donaldson, & Geddes, 1997)

Class IIb:

- Use by family members of high risk individuals (Level of Evidence C; Priori et al., 2000; Page, Hamdan, & McKenas, 1998)

Definitions:

Levels of Evidence:

A. Data derived from multiple randomized clinical trials or meta-analyses
B. Data derived from a single randomized trial or non-randomized studies
C. Consensus opinion of the experts

Class of Recommendation:

Class I: Conditions for which there is evidence or general agreement that a given procedure or treatment is useful and effective

Class II: Conditions for which there is conflicting evidence or a divergence of opinion about the usefulness/efficacy of a procedure or treatment

Class IIa: weight of evidence /opinion is in favour of usefulness/efficacy

Class IIb: usefulness/efficacy is less well established by evidence/opinion

Class III: Conditions for which there is evidence and/ or general agreement that the procedure/ treatment is not useful/effective and in some cases may be harmful

CLINICAL ALGORITHM(S)

Algorithms are provided for basic life support and advanced cardiac life support.

EVIDENCE SUPPORTING THE RECOMMENDATIONS

REFERENCES SUPPORTING THE RECOMMENDATIONS

References open in a new window

TYPE OF EVIDENCE SUPPORTING THE RECOMMENDATIONS

The level of evidence and class of recommendation is given for selected recommendations (see "Major Recommendations" field).

BENEFITS/HARMS OF IMPLEMENTING THE GUIDELINE RECOMMENDATIONS

POTENTIAL BENEFITS

In general, risk stratification may identify high risk patients, which may reduce morbidity and mortality from sudden cardiac death.

Subgroups Most Likely to Benefit:

Patients with a history of congestive heart failure or depressed left ventricular function tend to show the greatest benefit in mortality reduction from beta-blockers.

POTENTIAL HARMS

- Sodium channel blockers
    - Prophylactic administration of sodium channel blockers is associated with an increased risk of all-cause mortality solely based on a pro-arrhythmic effect of this therapy.
- Amiodarone
    - Syncope and increased non-sudden death rate
- d,l sotalol
    - Can cause torsades de pointes tachycardia

QUALIFYING STATEMENTS

QUALIFYING STATEMENTS

- It should be reinforced that these recommendations cannot be intended as comprehensive 'guidelines for treatment' in those conditions such as myocardial infarction and failure in which sudden cardiac death is only one of the causes of death. In these conditions the objective of treatment has to be 'total mortality'. It is recognized that these guidelines do not consider the global management of all aspects of pathophysiological states associated with sudden cardiac death. However, interventions that are known to prevent sudden cardiac death are identified and emphasized.
- At the end of each section of the document, recommendations for risk stratification and for prevention of sudden cardiac death are provided. When reading the document it is important to remember that the recommendations are not for the comprehensive management of patients with any given condition but are only intended to highlight the value of different risk stratifiers to identify patients at risk of sudden cardiac death and to rank the effectiveness of different interventions in preventing sudden cardiac death. When treating patients this advice should be placed in the appropriate clinical context.

- The use of the terms 'primary' and 'secondary' prevention within the guideline is unconventional as it refers to patients with/without a history of sustained ventricular arrhythmia/ventricular fibrillation.

IMPLEMENTATION OF THE GUIDELINE

DESCRIPTION OF IMPLEMENTATION STRATEGY

An implementation strategy was not provided.

INSTITUTE OF MEDICINE (IOM) NATIONAL HEALTHCARE QUALITY REPORT CATEGORIES

IOM CARE NEED

Getting Better
Living with Illness
Staying Healthy

IOM DOMAIN

Effectiveness

IDENTIFYING INFORMATION AND AVAILABILITY

BIBLIOGRAPHIC SOURCE(S)

Priori SG, Aliot E, Blomstrom-Lundqvist C, Bossaert L, Breithardt G, Brugada P, Camm AJ, Cappato R, Cobbe SM, Di Mario C, Maron BJ, McKenna WJ, Pedersen AK, Ravens U, Schwartz PJ, Trusz-Gluza M, Vardas P, Wellens HJ, Zipes DP. Task Force on Sudden Cardiac Death of the European Society of Cardiology. Eur Heart J 2001 Aug;22(16):1374-450. [586 references]

ADAPTATION

Not applicable: The guideline was not adapted from another source.

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GUIDELINE DEVELOPER(S)

European Society of Cardiology - Medical Specialty Society

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GUIDELINE COMMITTEE

Task Force on Sudden Cardiac Death

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FINANCIAL DISCLOSURES/CONFLICTS OF INTEREST

Not stated

ENDORSER(S)

Danish Society of Cardiology - Medical Specialty Society
Estonian Cardiac Society - Medical Specialty Society
Italian Federation of Cardiology - Medical Specialty Society
Latvian Society of Cardiology - Medical Specialty Society
Lebanese Society of Cardiology - Medical Specialty Society
Polish Cardiac Society - Medical Specialty Society
Portuguese Society of Cardiology - Medical Specialty Society
Slovak Society of Cardiology - Medical Specialty Society
Society of Cardiology of the Russian Federation - Medical Specialty Society
Spain National Society of Cardiology - Medical Specialty Society
Turkish Society of Cardiology - Medical Specialty Society

GUIDELINE STATUS

Please note: This guideline has been updated. The National Guideline Clearinghouse will update this summary when the updated version of the full text document is published.

GUIDELINE AVAILABILITY

Electronic copies: Available in Portable Document Format (PDF) from the European Society of Cardiology (ESC) Web site.

Print copies: Available from Elsevier Publishers Ltd. 32 Jamestown Road, London, NW1 7BY, United Kingdom. Tel +44.207.424.4200/ Tel: +44 207 424 4389; Fax: +44 207 424 4433; e-mail: gr.davies@elsevier.com; Web site: www.escardiocontent.org.

AVAILABILITY OF COMPANION DOCUMENTS

The following are available:

- Recommendations for Task Force creation and report production. Sophia Antipolis (France): European Society of Cardiology, 2002.

Electronic copies: Available in Portable Document Format (PDF) from the European Society of Cardiology (ESC) Web site.

- Task Force on Sudden Cardiac Death, European Society of Cardiology. Summary of recommendations. Europace 2002 4:3-18.

  Electronic copies: Available in Portable Document Format (PDF) from the European Society of Cardiology (ESC) Web site.

- Guidelines on prevention of sudden cardiac death. Pocket guidelines. Sophia Antipolis (France): European Society of Cardiology, 2003.

  Electronic copies: An order form for ESC pocket guidelines is available in Portable Document Format (PDF) from the European Society of Cardiology (ESC) Web site.

- Guidelines on prevention of sudden cardiac death. Educational slides (Microsoft PowerPoint). Sophia Antipolis (France): European Society of Cardiology, 2003.

  Electronic copies: An order form for ESC pocket guidelines is available in Portable Document Format (PDF) from the European Society of Cardiology (ESC) Web site.

PATIENT RESOURCES

None available

NGC STATUS

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